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A CASE OF CHRONIC INTERNAL HYDROCEPHALUS IN A YOUTH.*

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Following is a case of chronic internal hydrocephalus occurring in a youth ten years after fracture of the skull. After death were found organized basal meningitis and focal sclerosis of the right anterior choroid artery:

An Irish boy of seventeen entered the Boston City Hospital, August 30, 1902, with symptoms and signs of heightened intracranial tension.

Ten years before he had suffered a compound fracture of the skull to the left of the vertex, from which he was perfectly recovered by the end of four months. Seven and a half years before the present illness, he suffered a somewhat severe contusion of the occiput by a kick by a horse; there was considerable swelling, apparently with infection; but he recovered in the course of time with repeated dressings, all the while an out-patient. He had recently, until within seven months, worked in a harness shop. Habits were excellent; the family history irrelevant. Scarlet fever and malaria were the only acute diseases remembered.

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During the seven months preceding his entrance, he had developed a rather characteristic set of symptoms:

1. Vertigo.
2. Unconsciousness at irregular intervals in attacks lasting two minutes. There was a peculiar aura, consisting of numb feelings about the eyes and in the tongue. Then followed agitation of limbs, gradually shared by the whole body. The tongue was not bitten in the attacks.
3. Projectile vomiting, usually after breakfast, occasionally after other meals, without retching or nausea. This had ceased three months before entrance.
4. Headache, general and nearly constant for the seven months.
5. Blindness, absolute for the last nine weeks. The "mist" had begun three months before in the left eye. When examined by one of the writers (W. F. R.) at the Massachusetts Charitable Eye and Ear Infirmary, five months before entrance at the City Hospital, there were already well marked choked discs, and the symptoms already strongly suggested brain tumor.

Physical examination showed no paralyses or sensory disturbance, except bilateral loss of smell and blindness. The tongue was protruded in the middle line. Movements of the face were symmetrical. The reflexes were scarcely affected: the left knee-jerk, absent; Babinski reflex and ankle clonus, absent. These phenomena, with ataxia, both standing and walking, pointed to lesion in the cerebellum (Dr. W. N. Bullard).

The operation was performed by Dr. J. C. Munro six days after entrance. Upon trephining the right occipital bone, there was no pulsation in the dura, and there was considerable hernia cerebelli, without evidence of new growth or other lesion. The right ventricle was tapped through a second trephine-opening, and there was a profuse discharge of several ounces of clear ventricular fluid under pressure. The pulsation then returned.

After the operation the patient was so far improved that he lay quietly for some days, with some pain at the site of the incisions, but without general headache. There was a slight improvement in vision, so that he could now distinguish light with the right eye. The knee-jerk came back upon the right side, but never became so marked as upon the left side. There was some ataxia in the movement of the hands.

Seventeen days later his temperature rose to 105° F., and he vomited several times. He had a clonic spasm of the right arm at intervals.

The right ventricle was again tapped, and considerable fluid was withdrawn under pressure. Pulsation of the dura was brought about once more; but the boy never regained consciousness and died the next morning, with the intracranial pressure unabated.

We may here express our obligations to Dr. W. N. Bullard, Dr. J. C. Munro, and the house-officers of the First Surgical Service for access to the clinical records.

Clinical Summary.—History of the head injuries, with perfect recovery: vertigo, attacks resembling Jacksonian epilepsy, projectile vomiting, headache, blindness, ataxia, loss of right knee-jerk, increase of left knee-jerk—the syndrome occupying seven months. Exploratory operation upon right posterior fossa cerebri and tapping of right ventricle. Some relief of symptoms. Tapping of ventricle a second time. Death eighteen days after initial operation. Duration of fatal illness, less than eight months.

Permission was given to open the head only. The examination, nineteen hours after death, was made by one of us (E. E. S.), during the service of Dr. F. B. Mallory, to whom we are indebted for aid in the report. Following is a description of findings:

Well developed, well nourished white boy. Rigidity of extremities and of arrectores pilorum. Lividity slight. Pupils dilated, left somewhat less than right. Operative incisions.

Pericranium adherent over nickel-sized area in the left frontal region. Inner table shows no sign of lesion at this point. Cranium of the usual thickness. No sign of fracture at the base.

Dura and cranium interadhere in parietal regions. There is firm interadhesion of tissues both within and without the cerebellar window. Sinuses filled with fluid blood.

Pia lifted from sulci in the vertex region by a clear fluid containing a few fibrinous flocculi. Pacchionian granulations extensively developed, as in old age. The pia at the base is quite regularly overgrown, especially about the olfactory nerves and the optic chiasm and around the vessels of the circle of Willis, which are normal. The overgrowth looks thicker than it actually is from the extreme edema. The overgrowth is of closest texture in the fossæ between medulla and cerebellum. The areolar tissue of the Sylvian fossæ is normal except for edema. The pia is everywhere edematous, but nowhere evidently thickened. The pial veins of the left side are more markedly injected than those of the right. The pia beneath the right hemisphere of the cerebellum is puffed out with perhaps fifty cubic centimeters of clear fluid. No evidence of tuberculosis.

Convolutions show marked flattening; both hemispheres are still under a degree of tension. Substance everywhere apparently normal, except where changes have followed the operations: there is very little injection in the track of the paracentesis; the right cerebellum is well dismantled, presenting a gray-red, thready, grumous mass, in which there are no naked-eye evidences of neoplasm or other lesion not caused by operation.

The ventricles contain ten to fifteen cubic centimeters of fluid, on the left side clear, on the right clear and red from the leaking of blood. Vessels in ventricle floor are equally injected on the two sides. Choroid plexuses on both sides exhibit several opaque yellowish masses, one to two millimeters in diameter. The upper surface of the right choroid plexus is overgrown with connective tissues, so that the fornix cannot be stripped forward without ripping out bits of brain substance. The right anterior choroid artery shows a well-marked yellowish patch of arteriosclerosis three millimeters in diameter, four centimeters from the origin of the artery, two-thirds occlusive.

Anatomical Summary.—Chronic leptomeningitis with organization, especially well-marked at the base, about the superficial origins of nerves I. and II., and between medulla and hemispheres of cerebellum. Focal sclerosis of right anterior choroid artery. Chronic inflammatory changes with organization in right choroid plexus. Psammomata of both choroid plexuses. Epicerebral edema. Internal hydrocephalus.

The foregoing is a bit of evidence in the case against idiopathic internal hydrocephalus.

It is possible to hold that internal hydrocephalus is, like external hydrocephalus, never a genuine entity.[1] Acute internal hydrocephalus[2] usually proves to be one aspect in the picture of a meningitis, of cerebral edema from fracture of the skull, of commotio cerebri, of alcoholism, of renal or cardiorenal disease. Those cases of internal hydrocephalus, classed acute, that have been likened to angioneurotic edema of the subcutaneous tissues,[3] and the cases of so-called serous apoplexy[4] have the best title to the term entity. These cases are probably best reckoned as chronic with exacerbations,[5] [6] or confessed as obscure.

It is less easy to dispose of chronic internal hydrocephalus as in all cases a partial phenomenon. Chronic internal hydrocephalus is systematically divided into the congenital and the acquired.[7] It would probably be better to distinguish the form running its course before union of sutures from a form, less well defined, occurring after sutural union.

The factors in the form of chronic internal hydrocephalus before union of sutures are uncertain, may involve a basal meningitis,[8] a dysostose cleidocrânienne,[9] or both, with an active secretion of lymph through the ependyma. A developmental factor is admitted or quite narrowly excluded.

The form of chronic internal hydrocephalus occurring after closure of sutures is usually regarded as arising in an exudative process, as the effect of stasis, as angioneurotic, or as forming *e vacuo*.[10] Of these, the cases *e vacuo* (in general paresis, softenings, and old age) may be set aside (in the group with external hydrocephalus, which they often accompany) as without doubt partial phenomena. The part assigned to an angioneurosis must be looked on as a more or less clever hypothesis, which so far explains nothing.

As possible entities there remain, among cases of chronic internal hydrocephalus setting in after closure of sutures, two accepted (or classified) forms: a form due to exudative processes and a transudative form with venous stasis. There is doubt whether venous stasis alone will occasion internal hydrocephalus. As to the inflammatory form, more and more cases are reported yearly which are putting the condition upon a satisfactory structural basis.[11]

Since the seventeen cases collected by Schulze,[12] the following cases of chronic internal hydrocephalus in adults, of interest on this point, have been reported:

1. Case of F. Taylor, 1897.—Boy of sixteen, with history of convulsions, bronchitis, and scarlet fever as a child. Symptoms referable to hydrocephalus extending two years, or possibly eighteen months, before death. Little characteristic in history beyond headache, vomiting and *tâche cérébrale.* Autopsy: Dilatation of all ventricles.[13]

2. Case of A. Heidenhain, 1899.—Male of twenty-three. Past history irrelevant. After exposure, headache, followed in three days by arching of neck, with head down and shoulders forward, lax extremities, heavy gait and slowness of speech. Later, in a course of several weeks, attacks of mania, with lowered temperature and pulse, lasting an hour. Autopsy: Not remarkable, excepting in a hydrocephalus involving all the ventricles without other lesion.[14]

3. Case of Burr and McCarthy, 1900.—Male of thirty-three. Fever, bradycardia, neck-stiffness, headache, stupor, hallucinations. Improvement after three weeks, with preservation of psychical changes. Recurrence of fever and meningitic symptoms

after one month. Death in a second recurrence. Papillæ normal. Knee-reflexes, at first absent, recurred just before death. Almost complete deafness. Autopsy: Hydrocephalus, ependymal proliferation of subependymal glia, perivascular infiltration beneath ependyma, old and fresh inflammation in choroid plexus. Similar changes without hydrocephalus were caused experimentally in cats by the injection into ventricles of various substances.[15]

4. Case of Spiller, 1902.—Male of fourteen and two-thirds years. Loss of power to walk with asymmetrical contractures and ultimate confinement to bed four years. Dilatation of right lateral ventricle from partial closure of foramen of Munro (inflammatory possibly tuberculous).[16]

5. Case of Spiller, 1902.—Male of nineteen. Headache with vomiting once or twice a week since childhood; six months' exophthalmos. Optic neuritis. Knee-reflexes increased. Death sudden. Autopsy: Cerebral ventricles dilated, fourth ventricle normal.[17]

6. Case of Grober, 1903.—Woman of twenty-four. History of malaise. No fever, slowing of pulse, or symptoms of previous meningitis. Case reported on account of focal asymmetrical symptoms, spasm in right facial region, hypesthesia in right supraorbital region, left facial paresis, left hypoglossus paresis, increase of right knee-reflex. Grober calls the anatomical finding *Hydrocephalus Acutus Internus.*[18]

7. Case of Gerhardt, 1903.—Male of twenty-two. Nine months before death, occipital pain, vomiting, vertigo, photophobia, with recovery after eight days. Seven months before death, recurrence. A second recurrence was apparently benefited by mercurial inunction (disappearance of choked disc). A third recurrence was characterized by apoplectiform attacks, with transient hemiplegia, once with double vision, without lasting focal symptoms. Autopsy: Hydrocephalus, ependymitis of fourth ventricle, obliteration of foramen of Majendie, fibrous thickening and cyst-formation of choroid plexus of fourth ventricle.[19]

Such are cases in adults which form the bulk of evidence that chronic internal hydrocephalus is an entity. The condition is scarcely entitled to the name, from mere obscurity of origin. The usual finding is chronic inflammation of plexuses, about foramina, or in meninges. There is a priori doubt whether chronic inflammatory changes will alone occasion hydrocephalus.[20] If they, however, will, then the supporter of hydrocephalus as entity must seek to establish chronic inflammation as entity. The work of Joslin in tracing the remote effects of epidemic cerebrospinal meningitis in producing internal hydrocephalus is perhaps the best example of work on the line of resolving these phenomena.[21]

A possible interpretation of the present case is to suppose lowered pressure in the choroid area, consequent upon focal sclerosis in the anterior choroid artery, with effusion of fluid, as in infarction. That such sclerosis, not infrequent in old age, should not more often bring about internal hydrocephalus, is due to the absence of chronic changes at the lymph-outlet (foramen of Majendie or proximally).

A somewhat similar case is the often-quoted case of Newman (1882), of thrombosis of the vena magna Galeni.[22] Trendelenburg mentions, as a pathological curiosity, a case of tuberculosis effecting pressure on the vein of Galen, venous congestion, and ventricular effusion.[23] There is little doubt that tumor masses or cicatrices of various origin could be appropriately set to bring about ventricular effusion and to prevent its escape.

Further cases may throw light on the part possibly played by vascular lesions of the anterior choroid arteries in setting up internal hydrocephalus.

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[1] Pilcz. "Behandlung des Hydrocephalus," Centralbl., f. d. Grenzgebiete der Medizin und Chirurgie, S. 681, 1899.

[2] Schulze. "Krankheiten der Hirnhäute und Hydrocephalie," Nothnagel's Specielle Pathol. und Therapie, IX. Band, III Theil, Wien, 1901.

[3] Quincke. "Meningitis serosa," Volkmann's Vorträge, 1893, Nr. 67; also Zeitschr. f. Nervenheilk, IX, 1896, and Berliner klin. Wochenschr., No. 39, 1891.

[4] Heidenhain. "Hydrocephalus acutus acquisitus (idiopathicus)," Berliner klin. Wochenschr., Nr. 49, 1899.

[5] Schulze. Loc. cit., S. 235.

[6] Prince. "Idiopathic Internal Hydrocephalus," p. 502, Journ. Nerv. and Ment. Dis., 1897. . .

[7] Heubner, in Eulenburg's Realencyclopædie, 1887.

[8] Lees and Barlow. "Simple Meningitis in Children," Allbutt's System of Medicine, Vol. VII, p. 492, 18.

[9] Marie et Santon. "Le Dysostose Cleidocrânienne Hereditaire," Bull, et Mem. Soc. med. d. Hop. de Paris, S., XV, p. 436, 1898.

[10] Heubner. Loc. cit.

[11] Joslin. "Internal Hydrocephalus following Cerebrospinal Meningitis," Am. Journ. Med. Sci., Oct., 1900.

[12] Schulze. Loc. cit., S. 230-236.

[13] Taylor, F. "A Case of Chronic Internal Hydrocephalus Fatal at Sixteen," Tr. Clin. Soc., London, XXX, p. 175-9, 1896-7.

[14] Heidenhain. Loc. cit.

[15] Burr and McCarthy. "Acute Internal Hydrocephalus," Journ. Exper. Med., October, 1900.

[16] Spiller. "Two Cases of Partial Internal Hydrocephalus from Closure of the Interventricular Passages," Am. Journ. Med. Sci., p. 44, 1902.

[17] Spiller. Supra.

[18] Grober. "Herdsymptome bei Hydrocephalus acutus internus der Erwachsenen," Mitt. a. d. Grenzgeb. d. Med. u. Chir., XI, 30-38, 1903.

[19] Gerhardt. "Drei Fälle von Hydrocephalus bei Erwachsenen," Wanderversammlung, Baden-Baden, 1903.

[20] Burr and McCarthy. Loc. cit.

[21] Joslin. Loc. cit.

[22] Newman. Glasgow Med. Journ., Sept., 1882.

[23] Trendelenburg. "Hydrocephalus," Internat. Clin., 8 S., III, p. 209,